339 Cystic fibrosis-related diabetes (CFRD) in the Czech Republic

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Data from multicentric studies demonstrate worsening of nutritional status, lung function and shortening of lifespan in CFRD patients. A cross sectional study of CF patients from Czech Republic (n = 471, 64 CFRD, 15 impaired glucose tolerance IGT) corresponds with data from literature. After 23 years of age the nutritional status and lung functions are significantly worse in the CFRD/IGT group (BMI −0.9±SD vs −0.2±1.2 SD, $p = 0.005$; FEV_1 51±22.9% pv vs 66.9±24.2% pv, $p < 0.01$). We compared these results with prospective pair study of 29 insulinopenic patients diagnosed by regular screening (oGTT) in the years 1997–2002. Controls were sex/age matched to cases. In this study we did not prove significant difference between the group treated by insulin and control group during the follow up period (range 5–9.6). At the last examination the median age of the patients was 23.4 years, FEV_1 in cases 62.3±25.4 vs 60.9±24.4 in controls. During the study in 4 controls insulinopenia was detected. In treated cases 4 females died, in controls 3 females and 3 males were deceased (one with insulinopenia). Cross sectional data in patients older than 23 years show adverse influence of CFRD on nutritional status and pulmonary function. Our prospective pair study of early detected and treated children did not prove this adverse effect at least after 5 years of treatment. We suppose that the worsening could be caused not only by long lasting hyperglycaemia, but especially by insulinopenia, insufficient management of CFRD and/or non-compliance.

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340 Glucose tolerance changes in cystic fibrosis (CF) patients during a 10 years period

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We evaluated impaired glucose tolerance and its variations during a 10 years period in 2 groups of CF patients:13 adults (10 F, mean age 30±3, range 25–36 yrs); 21 adolescents (8 F, mean age 20±3, range 14–24 yrs). At baseline (time A) and 10 years after (time B) we assessed the intravenous glucose tolerance test (IVGTT), body mass index (BMI, %) and forced expiratory volume at first sec (FEV_1, % pred.). At the time B the patients underwent oral glucose tolerance test (OGTT). Based on the first phase insulin response (FPIR) during the IVGTT, the patients were classified as normal (FPIR between 3–97º centile) and with insulin deficiency (FPIR below 3º centile). The OGTT was classified according to the CF international criteria. The FPIR values (mean± SD) were 100± 69 and 67±48 in adults and 68±43 and 61±43 in adolescents at time A and time B, respectively. No significant relationships were found between FPIR changes and age, BMI, FEV_1 changes in 10 years period. In the adult group 54% and 69% showed insulin deficiency at time A and time B, respectively. In adults with reduced insulin levels, OGTT revealed 2 diabetes.

In adolescent group 67% and 76% showed insulin deficiency at time A and time B, respectively. In young patients with reduced insulin levels, OGTT revealed 2 diabetes.

Our results confirm that insulin deficiency is very common in both adult and adolescent CF patients. In addition, our study did not find any significant changes in insulin levels through the time and no significant relationships between insulin secretion, nutritional status and pulmonary function.

341 Demographics of glucose metabolism and gender differences in cystic fibrosis

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Background: In this study we investigated the prevalence of cystic fibrosis-related diabetes (CFRD) in a Dutch CF centre for children and adults. Further the relationship between CFRD and several clinical parameters was studied.

Methods: All exocrine pancreatic insufficient CF patients older than 10 years underwent their yearly oral glucose tolerance test. All patients were classified as having either normal glucose tolerance (NGT), impaired glucose tolerance (IGT) or CFRD. Associations with the following parameters were studied: age, gender, BMI, mutations, FEV1 % predicted, FVC % predicted, chronic pseudomonas infections, *Burkholderia cepacia* infections, liver disease, ABPA, and hospitalization.

Results: In the total study population prevalence of IGT and CFRD is respectively 16% and 31%. After exclusion of all pancreas sufficient patients, prevalence of CFRD rises up to 40% and in patients over 40 years even up to 52%. In comparison with patients with a NGT, CFRD patients are older ($p = 0.022$), have worse pulmonary functions ($p < 0.001$) and stay longer in the hospital ($p = 0.002$). There is a tendency in CFRD patients towards more chronic pseudomonas infections and more ΔF508 mutations. Women develop CFRD on a significantly younger age than men (respectively 19.5 and 27.5 years, $p = 0.008$), though there is no difference in total prevalence.

Conclusion: Prevalence of CFRD is increasing in CF populations and is strongly associated with worse clinical status of the patients. Women develop CFRD on a younger age. This knowledge is important for understanding the pathophysiology and clinical importance of CFRD.

342 A review of CF-related diabetes (CFRD) management – a patients perspective

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Introduction: The UK CF Trust Diabetes working group 2004 highlighted a need for an annual review of CFRD and that monitoring and treatment is vital in preventing decline in weight and lung function. In 2006, the CF service based in Ninewells Hospital, Dundee had 40 adult patients of which 15 had developed CFRD. These patients attended a diabetes and CF clinic several times per annum. It was hypothesised that treatment and outcome measures could be improved if a joint service was provided. Patient views were sought on this and a new model of care adopted.

Method: The 15 patients with CFRD completed a questionnaire based on their experiences of attending both a diabetes and CF clinic.

Results: 14 questionnaires were completed and the main points extrapolated were:

1. 13/14 of patients had attended a diabetes clinic within the last year in a variety of locations within Tayside. Missed appointments were due to time pressures with other clinic/work/social commitments.
2. Patients perceived that the diabetes nurse and review of insulin treatment was the most helpful in clinic.
3. 11/14 would prefer to be seen as a joint review in CF clinic and none wanted to be see at the hospital diabetes clinic.

Following this, the diabetes specialist nurse attended clinic to provide support and advice for both patients and staff.

Conclusion: Whilst a joint annual review with CF consultant and Diabetologist is ideal, it is not always practical with small numbers. The presence of a Diabetes nurse in CF clinic improved both the patients understanding and control of diabetes and provided a more integrated service. Hospital visits were reduced whilst still completing annual health checks. There continues to be input on a regular basis with support from the Diabetologist. Practice will be reviewed in 2008 to look at key areas for further development.